Timing of Surgery for Cervical Spine Anomalies

Ilkka Helenius, MD, PhD
Professor and Chairman
Department of Pediatric Orthopedic Surgery
University of Turku and Turku University Hospital, Finland
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Pathology of Cervical Spine

- **Instability**
  - Occipitocervical (Down)
  - Atlantoaxial (Os odontoideum)
  - Subaxial (Klippel-Feil, Diastrophic dysplasia, Larsen syndrome)

- **Deformity**
  - Cervical kyphosis (Congenital, Neurofibromatosis)

- **Spinal stenosis**
  - Achondroplasia
Cervical Spine Instability

- **Occipitocervical instability (C0/C1)**
  - Basilar invagination: Tip of dens above level of foramen magnum (McRae line) (Chamberlain WE. Yale J Biol Med 1939;11:487-96)
  - Instability (Down, 22q11.2 Deletion syndrome): >1 mm of translation at the occipitoatlantal articulation (measured from the anterior surface of the occipital condyles to the posterior surface of the anterior arch of the atlas) (Wiesel SW. Spine 1979;4:187-91.)

- **Atlantoaxial instability (C1/C2)**
  - Atlantoaxial distance ≥5 mm (Locke GR, AJR 1966;97:135-40)
  - Space available for cord (SAC) < 13mm (Spierings and Braakman, JBJS Br 1982;64-B:422-8)
  - Os odontoideum, skeletal dysplasias such as spondyloepiphyseal dysplasia

- **Subaxial instability**
  - Vertebral body hypoplasia (congenital, diastrophic dysplasia)
  - Klippel-Feil with block vertebrae
  - Vertebral dysplasia and erosion (Neurofibromatosis)
  - Postlaminectomy (without instrumentation)
Flexion-Extension MR images

- Bony landmarks often difficult skeletal dysplasia
- Flexion-extension MR images diagnostic and safe (MacKenzie et al. JPO 2013)
- Protocol in children skeletal dysplasia
  - General anaesthesia or sedation. Radiologist and anesthesiologist available
  - T1+T2 sagittal, axial in neutral position (external auditory meatus at the level of shoulders)
  - If no cord compression / airway considerations continue with dynamic MRI
  - Flexion: Padding under occiput
  - Padding under shoulders
- Findings
  - Space available for cord, critical value less than 13 mm
  - Cord encroachment, signal changes
  - Soft tissue mass not shown in plain films or CT scan (Morquio) (Dede et al. JBJS 2013)
C Spine Instrumentation before 2 yrs

- Anatomic dimensions on CT more important than age (Lastikka et al. JCO 2017, in press)
- Occipitocervical fusion before the age of 2 years (Hale et al. Child Nerv Syst 2017;33:1253-69)
  - 10 infants with traumatic C0/C1 instability or C2 fx
  - 10 infants with congenital instability (3 with Down sdr, 3 with skeletal dysplasia, 4 with congenital anomaly
  - Complications: transversal sinus injury (1), worsening neurology (2), all fused
  - C1 lateral mass, C2 pedicle or translaminar screws can often be used; strut graft useful (rib, iliac)
Osteotomy classification of Cervical Spine

Hemivertebra Resection (C Spine)

• Traditional treatment for C spine hemi: Posterior in situ with/o instrumentation (Hensinger RN. Clin Orthop 1991)

• 1st description of cervical hemi resection by Deburge and Briard JBJS 1981 (C7, four staged surgery)

• Above C7 (in the vertebral artery area) described by Harms and Ruf (Spine 2005)
  – Combined posteroanterior or posteroanteroposterior approach
  – Three children, one developed C5 nerve palsy

• Harms and Ruf advocated early hemi resection to prevent compensatory curves

• Indications
  – Head tilt, prevention of compensatory curves if fully segmented hemi
Technical aspects of Hemivertebra Resection

- **Posterior exposure**
  - Hemilamina excision with facet joint above and below
  - Nerve root exposure, divisions of nerve root – vertebral artery connections
  - Resection of posterior wall for transverse foramen

- **Anterior exposure**
  - Disc above and below excised
  - Hemicorpus resected piecemiel
  - Transverse process resected with anterior wall of transverse foramen and exposure of vertebral artery
  - Anterior closure of resection with plate, if necessary posterior instrumentation with C2 pedicle and lateral mass screw

*Hemivertebra Resection in the Cervical Spine.*
Timing and Effects of C Spine Deformity

- Harms and Ruf advocated early hemi resection to prevent compensatory curves, age ranged 4.4 to 14.9 in three patients (Spine 2005).
- Anatomic dimensions need to be satisfactory for implant placement (Lastikka et al. JCO 2017)

Natural course of a cervical congenital scoliosis in a boy with hemivertebrae C3a and C7a at the left side. At the age of 2 years, the thoracic spine was completely straight (A). At the age of 14 years, there was a compensatory, now structural curve of 79[degrees] at the thoracic spine (B). Ruf et al. Spine 2005
Craniocervical Junction Hemi

- Described on adult patient (Ruf et al. Spine 2015)
- Visualization of basilar & vertebral arteries preoperatively
- Transoral resection + posterior compression
Cervicothoracic Junctio

Hemivertebra

- Present with head tilt, shoulder imbalance
- Typically outside vertebral artery area
- Recent case series of 18 children (aged 5.5 to 15.2 yrs) with hemis between C7 and T4 (Chen et al. Spine 2017, Jul 11, epub)
  - 58% scoliosis correction with hemivertebrectomy
  - Neck tilt: 20° → 11°
  - One Horner sdr, one radicular pain
  - 20% pedicle screw malposition risk
Klippel-Feil Instability

- Instability presents typically at adult age
- Contact sports denied
- Single level ACIF vs. posterior based instrumentation
Correction of Congenital Kyphosis >10yrs

- When neuro st + MR ok
- Postponing until rigid a+p fixation reliable is an option
Chondrodysplasia punctata

5-yr-old boy
Mid-cervical kyphosis and stenosis
Associated myelopathy
Custom-made collar since 1-yr-old
Neurosurgical plan: Anterior decompression via C4 corpectomy and plate with postop Halo immobilization
Immediate Postoperative MRI

Anterior corpectomy + biodegradable plate

No suitable halo body jacket available

Fixation failed and resulted into full tetraplegia + respiratory arrest (T2 sag MRI)
Posterior Decompression & C0-T2 Instr

Regained standing and eating but remains tetraparetic
Posterior decompression and C0-T4 fix

Non-union at 1.5 years postop, no anterior column support?

Combined approach revision with anterior support
At 4-yr Follow-Up Union

N. Pakkasjärvi; M. Mattila; V. Remes; I. Helenius; Scandinavian Journal of Surgery 2013;102:189-196.
Larsen’s syndrome

Connective tissue disorder

Congenital joint dislocations

Progressive cervical kyphosis and risk of neuro deficit with minor trauma (Johnston et al. JBJS 1996)

Anterior and posterior elements may become detached (Katz et al. JPO 2005)

Guidelines missing for treatment

Combined approach preferred (Madera et al. J Neurosurg Ped 2008)
Case from Literature

Spontaneous Correction in Diastrophic Dysplasia

- Kyphosis $<60^\circ \rightarrow$ resolving  
  (Remes et al., Spine 1999)
- Close follow-up 4 mths  
  - Custom made collar 4-6 kk>
- $>60^\circ$ AP fusion
- Fiberoscopic intubation for all procedures (tracheomalacia)

1-yr-old
4-yr-old
Implications in Early onset scoliosis treatment

EOS typically highly progressive in Diastrophic Dysplasia (Remes et al. Spine 2002)

Cervical kyphosis prevents Glisson traction during casting of scoliosis – Use underarm traction instead. Cervical kyphosis resolves by continued growth if less than 60 deg (Remes et al. Spine 1999)
Os Odontoideum

- **Os odontoideum**
  - Bone (os), tooth (odontoid), latin (Giacomini, Gior R Acad Med Torino 1886)
  - Lack of continuity between the odontoid process and the body of C2 (axis)
  - An independent ossicle with smooth cortical margins separated from a shorthened axis
  - Ossiculum terminale=non-union of secondary ossification center, not associated with C1/C2 instability

- **Clinical Presentation**
  - Asymptomatic
  - Neurologic deficits: Brainstem or spinal cord compression

- **Two main anatomical types**
  - Orthotopic: Ossicle associated with C1 anterior arch
  - Dystopic: Ossicle migrated towards clivus, functionally fused to the basion

- **Idiopathic and associated with syndromes**
  - Down syndrome, skeletal dysplasia

- **Atlantoaxial instability (C1/C2)**
  - Atlantoaxial distance (AAD) ≥5 mm (Locke GR, AJR 1966;97:135-40)
  - Anterior, posterior or combined
  - Measured between posterior border of anterior arch (C1) vs. Posterior border of body of axis
Radiographic findings

- Lack of continuity between the odontoid process and the body of C2 (axis)
  - An independent ossicle with smooth cortical margins separated from a shortened axis
  - Enlarged anterior arch of C1
  - Jigsaw sign = articulation between anterior arch and the os
- Two main anatomical types (Fielding et al. JBJS 1980)
  - Orthotopic: Ossicle associated with C1 anterior arch
  - Dystopic: Ossicle fused to the basion
- Atlantoaxial instability (C1/C2)
  - Atlantoaxial distance (AAD) ≥5 mm (Locke GR, AJR 1966;97:135-40)
  - Space available for cord (SAC) <13mm
- Canal encroachment
  - Bony
  - Reactive synovitis

Orthotopic

Dystopic

Reactive synovitis causing compression
Etiology

- **Embryology**
  - 4th occipital sclerotome (proatlas) → apical cap of dens and apical ligament
  - 1st spinal sclerotome (C1) → rest of dens
  - 2nd spinal sclerotome (C2) → axis body & arches

- **Congenital**
  - Fusion failure between the dens and body of atlas

- **Traumatic**
  - History of trauma common in os odontoideum

- **Etiology varies** (Sankar et al. Spine 2006)

- **Atlantoaxial instability (C1/C2)**
  - Atlantoaxial distance (AAD) ≥5 mm (Locke GR, AJR 1966;97:135-40)
  - Space available for cord (SAC) <13mm

- **Canal encroachment**
  - Bony
  - Reactive synovitis

Arvin et al. Neurosurgery 2010
Conservative treatment

- 20 patients treated conservatively (Spiering and Braakman JBJS Br 1982)
  - 15 patients without neurologic deficits, FU 6.5 years, none developed neurologic deficits
  - 4 patients with transient cord signs, no deterioration

- Minor trauma associated with
  - Spinal cord atrophy (Fielding et al. JBJS 1980)
  - Cerebellar infarction (Sasaki et al. Spine 2000)

- Minimum requirements for conservative treatment
  - Normal cord morphology
  - Minimum SAC >13 mm
  - 10% risk of myelopathy with SAC<13mm (Spiering and Braakman JBJS Br 1982)

- Conservative treatment includes
  - Stable, yearly flexion-extension radiographs
  - MR images every 5 years to prevent signal changes
  - No contact sports
  - Requires further investigations!
Preoperative considerations

- **C1/C2 instability**
  - AAD>4 mm, SAC<13mm
  - 10% risk of myelopathy with SAC<13mm (Fieding JBJS Br 1982)
- **C1-C2 vs. C0-C2 spinal fusion**
  - Down patients with dysplastic C0/C1 joints?
- **Preoperative imaging**
  - MR angiography: Vertebral arteries
  - CT: Bifid C1 posterior elements
  - Use of intraoperative navigation
- **Preoperative traction** (Hedequist et al. 2016)
- **Fixation points**
  - Occiput
  - C1 posterior elements, lateral mass
  - C2 posterior elements, pedicle
  - Transarticular screws

Dysplastic C0/C1 joints

Effect of preop traction on alignment (El-Barr et al. J Neurosurg Ped 2016)
Brooks-Jenkins wiring

- Spondyloepiphyseal dysplasia
- 4-yr-old girl
- Odontoid hypoplasia
  - AAD 8mm or SAC 9mm
Harms technique (C1 lateral mass, C2 pedicle)
Rigid fixation improves fusion rates

- 28 children with skeletal dysplasia and C1/C2 instability (Helenius ym. JBJS 2015)
- 14 non-rigid (mostly wiring), 14 rigid (Harms or hooks)
- Risk of non-union higher in non-rigid 6/14 vs. 0/14 (p=0.0057)
- Two vertebral artery lesions with attempted C2 pedicle screw placement
Author’s current practice

- Preoperative evaluation
  - MR: signal changes, cord morphology, anatomy of arteries (dominant vertebral artery?)

- Intraoperative traction
  - Halo + 1-2 kg traction

- Harms technique
  - C1 navigated + exposure of lateral mass (C2 sacrifice)
  - C2 free hand
  - Intraoperative O-arm to confirm reduction, implant placement
  - Avoid C0/C1 fusion

- Halo body jacket 2-3 months
- Custom made collar 3 months
- Contact sports not allowed

5-yr-old girl, head tilt, neck pain
Intraoperative traction

Make sure cervical alignment is acceptable with fluoroscopy
Implant placement
Intraoperative O-arm

Implant placement
Space available for cord
Alignment of os odontoideum of axis, avoid over/undercorrection
Conclusions

• Early surgery may prevent compensatory curves
• Anatomical dimensions need to be evaluated preoperatively
• If neurological status OK, postpone surgery until school age to allow stable, segmental, rigid fixation
• Ongoing study on Os Odontoideum by the Pediatric Cervical Spine Study Group
  – Idiopathic vs. non-idiopathic children
  – Conservative treatment
  – Operative treatment

Contact e-mail: ilkka.helenius@utu.fi
or jonathan.phillips@orlandohealth.com